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HYDROCEPHALUS AND ENDOCARDITIS IN A CONGENITAL SYPHILITIC MENTAL DEFECTIVE

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THE association of hydrocephalus with congenital syphilis is not as common as may be inferred from the relative frequency with which the disease attacks the central nervous system in young subjects. Jeans ¹ found that in a series of 214 congenitally syphilitic children, the cerebro-spinal fluid in 33 per cent. showed evidence of infection. Of 402 mentally defective children at Caterham Mental Hospital, 37 have been found to be the subjects of inherited syphilis, and of these the cerebro-spinal fluid in 12, or approximately one-third, gave abnormal reactions, such as positive Wassermann reaction, increase of cells and globulin, and colloidal gold curve ; hydrocephalus, however, only occurred in two of the 37, one of whom has since died, and forms the subject-matter of this paper. Of the other 365 non-syphilitic cases, there were 7 cases of hydrocephalus. In pre-Wassermann days Fournier ² reported 3 cases of hydrocephalus in his series of 233 congenital syphilitics. Since the introduction of the Wassermann reaction many workers have written on this subject.

Dean ³	(1909)	found 4	hydrocephalics	among 51	congenital syphilitics.
Gordon ⁴	(1913)	" 1	"	" 66	" "
Veeder ⁵	(1916)	" 2	"	" 100	" "
Woodall ⁶	(1928)	" 2	"	" 86	" "
Stewart ⁷	(1930)	" 5	"	" 190	" "

In the mode of progress of the disease, and the poor response to treatment, the case about to be described shows many features of interest.

A.M., male, was admitted to Caterham Mental Hospital on 19/1/23, aged 7 years, suffering from imbecility with hydrocephalus. He was unable to feed or to attend to himself.

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Family History.—The mother had had six pregnancies by her first husband, which terminated as follows :—

- (1) Miscarriage (five months).
- (2) Female, full term, died when four days old.
- (3) Female, still-born, seven months.
- (4) Miscarriage (five months).
- (5) Male (patient), full-term child. Hydrocephalus was noticed at end of one year.
- (6) Female, full-term, died aged 3, from meningitis.

The father had acquired syphilis four years before marriage and had only received trivial treatment. After marriage he developed an ulcer over his back for which he attended a general hospital, but refused specific treatment. He died suddenly at the age of 38. A hypertrophied heart and granular kidneys were stated to have been found post-mortem. Some years later the mother re-married, and on finding herself again pregnant she consulted her physician, and underwent a thorough course of anti-syphilitic treatment, with the result that a perfectly healthy child was born, the first that this unfortunate woman had had. This child, a girl, now 9 years old, is quite healthy ; she shows no signs of syphilis and is up to standard at school.

Personal History.—The patient's head was noticed to be getting bigger at the age of 12 months ; the fontanelles did not close for a long time. He was treated with pulv. hydrag. cum creta for over a year. He walked at the age of 4, had his tonsils removed at 5, and was stated to have had measles and whooping cough in infancy. On admission he had a rash over the buttocks, and old white scars over the trunk. His teeth were badly decayed, testes undescended.

In December, 1923, he developed varicella, June, 1924, rubella, December, 1924, ringworm of the scalp, April, 1925, double iritis, June, 1925, double interstitial keratitis. His blood Wassermann at the time was found to be strongly positive. He was then given a course of five intravenous injections of sulph-arsenol, grm. .06 to .12, over a period of five weeks. He also received pulv. hydrag. cum creta, grs. 1 daily. This appears to have had little effect on the course of the interstitial keratitis. At varying intervals up to August, 1929, he was given long courses of mercury in the form of grey powder. In

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August, 1929, when he came under my immediate care, his blood Wassermann was ++ (7 M.H.D.), his cerebro-spinal fluid negative. His gums were spongy and inflamed; he had some discharge from the eyes with steaminess of both corniæ. His pupils were inactive to light, his heart was of normal size with no murmurs, testes descended. In June, 1931, he had a seizure lasting three minutes, during which he was quite unconscious. In September, 1931, he had an attack of bacillary dysentery, lasting two weeks. His mental condition at the time showed signs of deterioration; he took little interest in his surroundings, and was quite unemployable. He showed no ataxia, and his speech was not of that drawling type usually associated with cases of juvenile general paralysis. About that time it was also noticed that he was becoming increasingly deaf. On 16/2/33 he developed a cellulitis of the left leg, followed by an endocarditis. He died on 9/3/33.

POST-MORTEM EXAMINATION—THIRTY-EIGHT HOURS AFTER DEATH

There were incisions over the lower part of the left leg and dorsum of the foot, from which some pus exuded. The underlying periosteum and bone were not affected.

Brain.—Weight, 62½ oz. Dura mater thickened and adherent to the calvarium in the right temporal region. Pia mater was much thickened and cloudy over all regions of the brain except the occipital lobes. There were cystic pockets of cerebro-spinal fluid and dense adhesions to the dura mater in many places, more especially over the left frontal lobe. The base of the brain was one mass of adhesions. The superficial blood vessels were numerous and thickened. The lateral ventricles, the foramina of Monro, and the third ventricle were all much dilated and contained clear cerebro-spinal fluid. The ependyma lining the ventricles was granular, and the choroid plexus atrophied. The beginning of the iter was much narrowed; the fourth ventricle beyond was of normal size. The skull was not thickened, and there was an area of rarefaction in the left parietal bone, close to the anterior fontanelle. The bridge of the nose was thickened and deformed, but there was no actual sinking in.

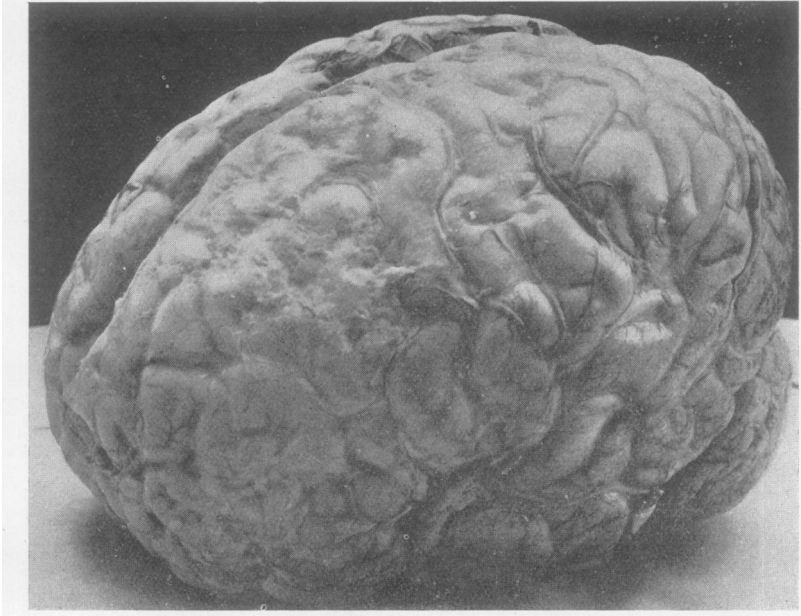
Lungs.—Right, 15 oz., left, 12½ oz. (Edema of both

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lower lobes. All the bronchi were thickened and when cut across oozed pus.

Heart.—10 oz. Myocardium firm, tricuspid valves thin and healthy, aortic valve cusps uniformly thickened, mitral valve thickened with many fairly recent vegetations close to the free border of both cusps.

Aorta.—Not dilated. The adventitia was adherent to the surrounding organs by bands of fibrous tissue. The intima showed many small patches of fibrosis.



Brain, showing thickened Pia mater with torn adhesions in the region of the upper part of the left psycho motor area.

Liver.—44 oz. Capsule not thickened, cut surface greasy and mottled.

Spleen.—6½ oz. Thickened capsule, firmly adherent to the stomach.

Kidneys.—Right, 4 oz., left, 4½ oz. Capsules thickened and somewhat adherent to the cortex. Pyramids and cortex well defined.

Stomach.—Many small submucous hæmorrhages.

HISTOLOGICAL EXAMINATION

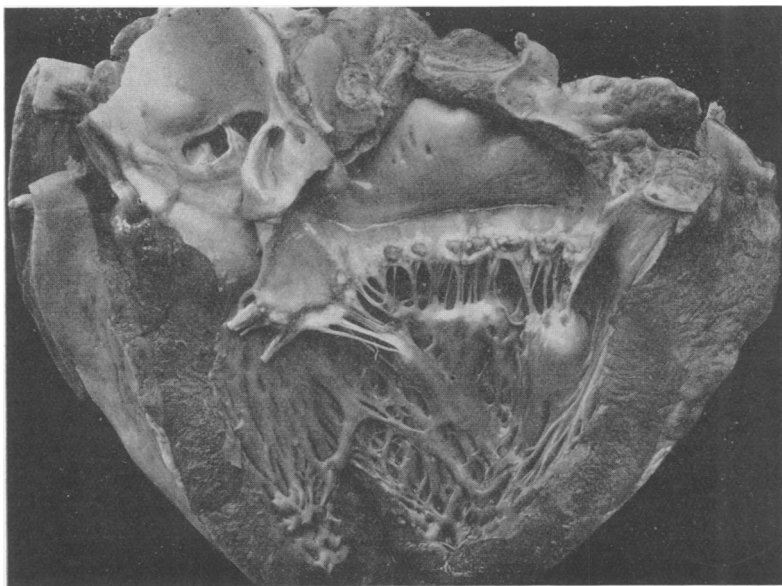
Brain.—Paraffin sections of the frontal cortex stained by hæmatoxylin and eosin showed lack of differentiation

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into cell layers. The pia mater was thickened, and some of the smaller blood vessels in the depths of the sulci were thrombosed. Their walls were much thickened, but there was no perivascular infiltration. There were very few nerve cells in the cortex, and these were mostly small and mis-shapen, with eccentric nuclei. There were numerous thickened blood vessels.

Paraffin sections of the cortex, stained by the Prussian blue method, failed to show any iron deposit in any of the neuroglial cells.

Liver.—Frozen sections stained by Scharlach R. showed



Heart, showing vegetations on the Mitral valve.

fatty degeneration of the parenchymal cells. Paraffin sections stained by hæmatoxylin and eosin showed dilated intralobular capillaries, and much increased fibrous tissue along Glisson's capsule. The bile ducts in this region were surrounded by whorls of fibrous tissue. In some places the portal vessels, which were all thickened, showed perivascular infiltration with small round cells. Close to the liver capsule there were many small scars.

Spleen.—The main feature was an increase of fibrous tissue.

Kidneys.—Showed thickening of Bowman's capsules.

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Heart.—Showed much increase of fibrous tissue between the muscle bundles ; the branches of the coronary arteries were much thickened.

Mitral Valve.—In the region of one of the vegetations showed almost complete fibrosis of the outgrowth.

Aorta.—Showed intimal thickening in places. The vasa vasorum in the adventitia were thickened, but no perivascular infiltration.

Thyroid.—The vesicles were normally filled with colloidal material, but there was an increase of inter-vesicular fibrous tissue.

Pituitary Body.—Showed a large number of dilated blood vessels in the anterior portion. The three types of cells were normally present.

Testes.—The intertubular fibrous tissue was very much increased, the blood vessels were much thickened, the cells of Leydig were plentiful and mostly situated in close relation to the blood vessels. The basement membrane of the tubules was thickened, cells of sertoli and spermatogonia were there, but little mitosis was present in the latter. Spermatocytes were few in number, and there were no spermatids or spermatozoa. The tubules were not dilated with secretion.

Epididymis.—There was much increase in fibrous tissue, which had in places much narrowed, and even obliterated the lumen of this part of the seminiferous channel.

Pieces of aorta, heart-valve, heart, liver, kidney, testes and epididymis, pituitary gland and cerebral cortex were stained by Jahnke's silver method for spirochaetes, but none were found.

Sections of liver and mitral valve were stained by Gram's method, but no Gram-positive organisms could be seen.

COMMENTS

The influence of untreated syphilis in the parents is well borne out in this case, where no healthy issue resulted from any of the six pregnancies. Though we cannot be quite certain, yet it seems probable that the effect of early treatment of the mother during her last pregnancy resulted in a healthy child.

In 1921, Hendry⁸ pointed out the good results to be expected from adequate anti-syphilitic treatment of pregnant syphilitic mothers. He mentioned a series of 43

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pregnancies in non-treated syphilitic mothers which ended as follows : 15 abortions, 17 still-births, 4 neo-natal deaths and only 7 living children. In 40 syphilitic mothers who received treatment during pregnancy, the results were : 2 abortions, 4 still-births, 5 neo-natal deaths and 29 living children. To 11 syphilitic mothers who were under treatment before pregnancy, 11 living children were born.

My experience, gained by personally interviewing the mothers of syphilitic mental defectives at Caterham Mental Hospital, is that very few of these women appear to avail themselves of the many excellent facilities for thorough treatment that exist in these days.

On the other hand, even prolonged anti-syphilitic treatment of the infected child unfortunately fails in many cases to affect the course of the disease, especially when such complications as interstitial keratitis supervene. In 1921, Veeder ⁹ stated that in spite of the fact that he had treated by various methods many of the 500 cases of congenital syphilis in St. Louis between 1912 and 1920, it was his feeling that when the whole thing was summed up, the results of treatment were most unsatisfactory. It was his belief that very little was to be gained by the treatment of the syphilitic child. He thought that the father and mother should receive treatment to prevent the birth of a child with hereditary syphilis. The course of the disease in the case above described shows how disappointing treatment can be.

The failure to find spirochoetes in the many organs examined calls for comment. Warthin ¹⁰ is of opinion that the spirochoete of syphilis quickly disappears from body tissues after the body has become cold. If this is so, it would offer an explanation, as the post-mortem was done thirty-eight hours after death.

The association of congenital syphilis with endocarditis is not common ; the presence of warty growths on the mitral valve cannot be said to be directly due to syphilis, as, according to Osler ¹¹ " a fresh, warty endocarditis due to syphilis is not recognised, though occasionally in persons dead of the disease this form is present, as is not uncommon in conditions of debility."

My thanks are due to Dr. J. E. McCartney of the Southern Group Laboratory for his helpful advice, and to Dr. T. Lindsay, Medical Superintendent of Caterham

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Mental Hospital, for kind permission to publish particulars of this case.

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